Coarctation of the Aorta

ANESTHETIC CONSIDERATIONS:
- Obstruction to systemic blood flow
- Increased LV afterload, HTN
- CHF in neonates and infants
- May be ductal-dependent as newborn – PGE_1 infusion to maintain ductus arteriosus patency
- Arrythmias
- SBE prophylaxis prior to dental or surgical procedures
- For surgery, considerations of aortic cross-clamping
- For ballooning, considerations of cardiac catheterization and remote location anesthesia

ANESTHETIC GOALS:
- Pre-operative optimization of LV dysfunction and heart failure if present
- Hemodynamic goals: low SVR, normal PVR, normal contractility, low HR, maintain coronary perfusion pressure
- Critical aortic coarctation and ductal-dependent blood flow: careful balance between systemic and pulmonary blood flow
- For surgery, be prepared to deal with HTN secondary to cross-clamp or post-operative paradoxical HTN
- For cardiac catheterization, be prepared to deal with the hemodynamic consequences of ballooning the aortic narrowing

HISTORY
- Clinical diagnosis, any previous surgical repair?
- **Functional status:** activity level, weight gain if infant, signs of low cardiac output or heart failure in the neonate including irritability, diaphoresis, cyanosis
  - Most adults with coarctation of the aorta are asymptomatic
    - May have a history of headache, dizziness, epistaxis, palpitations, leg claudication
- **Associated conditions and syndromes:**
  - Cardiac: PDA, bicuspid aortic valve, VSD, MS or MR, and various other types of left-sided obstructive lesions
  - Diffuse inherited angiopathy that predisposes patients with coarctation to cerebral aneurysm formation, aortic aneurysm development, and even aortic rupture - These medial aortic wall abnormalities appear to be part of a number of congenital cardiac anomalies and can involve the systemic and pulmonary arterial walls
  - Turner's syndrome, Williams syndrome, Alagille disease
  - VACTERL association
- **Medications:** inotropes, prostaglandins, afterload reducers- vasodilators, ace inhibitors, beta-blockers
- **Allergies, PMedHx, PSurgHx, PAnesthHx**

PHYSICAL
- **General:** weight, height, RR, baseline HR, systemic HTN, SaO_2, cyanosis, clubbing, FTT
- **Cardiac:** harsh systolic ejection murmur along LSB and in the back (especially over the area of the coarctation), brachial and femoral pulse volume (weak and delayed or absent femoral pulses), BP (sbp: arms>legs in postductal coarctation, dbp is same)
- **Signs of CHF:** elevated JVP, pedal edema, crackles, tachypnea, tachycardia, diaphoresis, hepatomegaly in infants
- **Respiratory:** signs of respiratory insufficiency (nasal flaring, increase RR, chest retractions, accessory muscle use)

INVESTIGATIONS
- **Labs:**
  - CBC (Hgb = 150-200 g/L for full term infant, at 2-3 months Hb = 90-100 due to physiologic anemia)
  - Coagulation studies- coagulopathy 2nd to low levels of vitamin K dependent factors
  - Lytes if medications dictate investigation
  - Renal function if associated syndromes (eg. renal anomalies with Turner's syndrome)
  - ABG if cyanosis or respiratory failure
- **ECG:** LVH, repolarization changes
- **CXR:** cardiomegaly, ascending aorta dilation, rib notching (late phenomenon only - yrs), increased collateral flow through the intercostal arteries causes symmetrical notching of the posterior third of the third through eighth ribs; the coarctation may be visible as an indentation of the aorta with prestenotic or poststenotic dilation of the aorta, producing the "reversed E," or "3," sign
- **Echo:** may visualize coarctation and arch anatomy, Doppler examination to estimate the transcoarctation pressure gradient (> 30 mmHg need surgery)
- **CT, MRI, and contrast aortography:** provide precise anatomic information regarding the location and length of the coarctation and the degree of collateral circulation
- **Heart catheterization:** if MRI unavailable and for coronary angiography when indicated

OPTIMIZATION
- Initial treatment:
  - Relieve CHF
  - Improve systemic perfusion – Maintain ductus arteriosus patency in severe or symptomatic neonatal coarctation with infusion of PGE_1 until surgical or transcatheter correction; low dose inotropic support may be required to optimize systemic perfusion
  - Improve or maintain pulmonary blood flow – Avoid hyperventilation, alkalosis, hyperoxia which decreases PVR and can increase pulmonary blood flow leading to systemic hypoperfusion; Aim for normal to mild hypercarbia
  - Mechanical ventilatory support with low FiO_2 may be used to maintain appropriate ventilation
- **Medications:** Children may be on sodium nitroprusside, labetalol (0.1-0.4 mg/kg q5-10 min or 0.25-1 mg/kg/h infusion), ACE inhibitors, etc. perioperatively, intraoperatively and/or postoperatively to control systemic hypertension during and after repair of aortic coarctation; Continue all meds except for diuretics and anticoagulants
- **Anxiolysis:** Midazolam 0.5 mg/kg IV / PR (maximum 20 mg), lorazepam 1-2 mg PO for adolescence
- SBE prophylaxis

ANESTHETIC OPTIONS
- For surgery: Regional eg. epidural is occasionally used for neonatal repair, GA

- For cardiac catheterization: Local and sedation, Regional, or GA (infants and small children frequently cannot tolerate the procedure under intravenous sedation and are more readily managed with GA)

ANESTHETIC SETUP

- **Drugs:** Standard emergency drugs, have inotropes drawn up and ready
- **Monitoring:** Standard CAS monitoring plus five-lead ECG, temperature measurement (rectal for cardiac cath), Foley catheter
    - Children undergoing repair of coarctation of the aorta should be monitored with a pulse oximeter on the right upper limb, because it may be the only reliable monitor during the repair; and both pre- and post-coarctation blood pressure cuffs should be placed; These two cuffs may be cycled, and the differential noted, before and after surgical correction
- **Lines:** Peripheral iv and arterial line in the right upper extremity to allow bp measurement during arterial cross clamping
    - Some have advocated an arterial line below the coarctation as well to measure perfusion pressure during cross clamping, but this may be very difficult in practice because femoral pulses are usually absent
    - In patients with left ventricular dysfunction, a central venous catheter may be desirable for pressure monitoring and inotropic support (Keep in mind that the left subclavian may be partially obstructed during the repair)
- Warming devices
- Have back-up rapid-response ECMO available (in case of cardiac arrest)

MANAGEMENT OF ANESTHESIA

- Depends on treatment: surgical repair or balloon stenting
- Surgical repair: for resting trans-coarctation pressure gradient > 30 mmHg or resting / exercise induced HTN

MANAGEMENT OF ANESTHESIA FOR SURGICAL REPAIR

- Surgery usually takes place through a left thoracotomy without the use of CPB
- The aorta is cross-clamped and the coarctation is repaired with an end-to-end anastomosis, patch aortoplasty, or subclavian patch

Induction:

- Maintain hemodynamic stability with careful induction, opioid + induction agent (etomidate in older children is good because of its cardiovascular stability) + muscle relaxant
- The lung will be retracted, and ventilation may be problematical; use a "snug" fitting ETT that has no leak

Maintenance:

- Hemodynamic goals:
    - Contractility- normal
    - Rate- slow hr
    - Rhythm- NSR
    - Afterload- normal to low SVR (but maintain coronary perfusion pressure and renal perfusion pressure during cross-clamp), normal PVR
    - Preload- adequate preload
- Aorta cross-clamping considerations:
    - Ensure adequate perfusion to the lower portion of the body during cross-clamping
        - MAP in the lower extremities should be at least 40 mm Hg to ensure adequate blood flow to the kidneys and spinal cord
        - If the systemic blood pressure cannot be maintained above this level, it may be necessary to use partial circulatory bypass
    - Propensity for systemic HTN during cross-clamping
        - Excessive increases in systolic blood pressure during cross-clamping of the aorta may adversely increase the work of the heart and make surgical repair more difficult
        - Use of volatile anesthetics is helpful for maintaining normal systemic blood pressures
        - If systemic hypertension persists, continuous intravenous infusions of nitroprusside should be considered
        - The disadvantages of lowering the systemic blood pressure to normal levels are excessively decreased perfusion pressure in the lower part of the body and inadequate blood flow to the kidneys and spinal cord
        - Some centers allow significant proximal hypertension (20% to 25% increase over baseline), based on evidence that vasodilator therapy may jeopardize distal perfusion and promote spinal cord ischemia
    - Risk of neurologic sequelae due to ischemia of the spinal cord (blood flow to the anterior spinal artery is augmented by radicular branches of the intercostal arteries and may be compromised during cross-clamping of the aorta for surgical resection of coarctation of the aorta)
        - Somatosensory evoked potentials are useful for monitoring spinal cord function and the adequacy of its blood flow during cross-clamping of the aorta; however, monitoring posterior (sensory) cord function does not ensure adequate blood flow to the anterior (motor) portion of the spinal cord
        - To reduce the chance of spinal cord damage, infants should be cooled to 34°C to 35°C before the cross clamp is applied and ventilated to normocarbia and upper limb blood pressure is maintained
        - Low-dose anticoagulation may also be applied
        - A short cross clamp time is also thought to be important
        - Although a collateral blood supply is present in older children, the spinal cord remains at risk during the cross clamping and the same precautions taken with neonates should also be taken with these children
- Release of aortic cross-clamping considerations:
    - Intravascular volume loading with 10 to 20 mL/kg of crystalloid is given just before removal of the clamp
    - Decrease anesthetic concentration
    - Give additional blood volume support until the blood pressure rises

Emergence:

- Early extubation is the norm
- Post-repair rebound hypertension due to heightened baroreceptor reactivity is common and often requires medical therapy
    - Ensure good analgesia
    - Institution of β-blockade with esmolol or labetalol during aortic cross-clamping can reduce post-repair HTN
        - Patients younger than 6 years of age should receive a loading dose of 250 to 500 μg/kg of esmolol, followed by an infusion of 250-750 μg/kg/min, depending on the blood pressure
    - Despite an esmolol infusion, 25% to 50% of patients have a blood pressure that is above the targeted range, requiring a second drug such as sodium nitroprusside
    - Other agents that may have a greater likelihood of achieving the targeted pressure include nitroglycerin and nicardipine
    - Propranolol is useful in older patients but can cause severe bradycardia in infants and young children

- Captopril or an alternative antihypertensive regimen is begun in the convalescent stage of recovery in those patients with persistent hypertension
- Many children, perhaps up to 30%, will go on to have long-term hypertension that will require therapy

MANAGEMENT OF ANESTHESIA FOR BALLOON STENTING

- Catheter techniques that include balloon angioplasty with and without stent implantation have been shown to be effective in relieving the stenotic aortic obstruction and normalizing blood pressure
  - This may be used as a primary therapy or to address residual or recurrent disease
  - Avoid surgical intervention, shortened hospital stay
  - Particularly beneficial to patients with recurrent coarctation, who are at a higher risk of operative intervention
  - However, a higher incidence of subsequent aortic aneurysm and recurrent coarctation than surgical resection

Induction:

- Anesthetic techniques used in these cases range from sedation and analgesia to general anesthesia
- Older, cooperative patients can be managed with iv sedation and analgesia with fentanyl, midazolam, propofol, and/or ketamine
- Infants and small children frequently cannot tolerate the procedure under iv sedation and are more readily managed with GA
  - If iv access is not present, inhalation induction with nitrous oxide, oxygen, and sevoflurane can be performed, and iv access is obtained after the patient is anesthetized
  - Alternatively, iv induction with thiopental, ketamine, etomidate, or propofol is performed
  - A nondepolarizing neuromuscular relaxant is administered for intubation
- In sick children in whom it may be preferable to use an iv induction, various options are available
  - In neonates with coarctation who are not ventilated before coming to the operating room, one approach is to administer fentanyl (2 to 3 μg/kg) to obtund the hypertensive response to intubation, followed by pancuronium to maintain CO by maintaining hr, and then very low dose (sedative dose) of sevoflurane or isoflurane for sedation/anesthesia
  - In older children, etomidate provides stable hemodynamics, although it does cause pain on injection
  - Ketamine is also widely used for iv induction in neonates and older children (maintains or increases bp, hr, and CO)

Maintenance:

- For GA, anesthesia is maintained with volatiles or TIVA using combinations of opioids, benzodiazepines, propofol, and ketamine
- Controlled ventilation avoids the increases in $Paco_2$ frequently seen with levels of sedation adequate to allow performance of this invasive procedure in pediatric patients
  - Minute ventilation and the respiratory rate are adjusted to maintain normal $Paco_2$ (to avoid alterations in PVR) based on analysis of arterial gases drawn by the cardiologist from the arterial catheter
  - The end-tidal CO_2 determination can be used to subsequently adjust ventilation, but it must be remembered that physiologic dead space is highly variable in these patients
- The potential hemodynamic behavior of the child depends on the nature of the lesion:
  - During balloon inflation, the circulation is blocked, and severe hypotension may result (and may not resolve immediately on deflation of the balloon)
    - Inotropic and antiarrhythmic therapy may be necessary
    - Preload may need to be optimized with iv fluid administration
  - A neonate with duct-dependent critical stenosis and little antegrade flow will tolerate balloon dilation well because there is little disruption of the cardiac output
  - Neonates and infants with less critical stenosis can suffer significant reductions in cardiac output when the balloon is inflated, especially if the ductus arteriosus is not patent
  - Older children tend to tolerate balloon valvuloplasty surprisingly well, with life-threatening hypotension being uncommon
- Close monitoring of these patients is required, especially in neonates, as deterioration can be rapid
  - These patients may be very sensitive to anesthetics, and hemodynamic instability may ensue
  - Repeated blood gas analysis is necessary because metabolic acidosis may be the initial sign of a low cardiac output state
  - Hypocalcemia and hypoglycemia may develop in neonates and require treatment
  - Hypothermia can be a problem in young patients, especially those under general anesthesia
  - Blood loss during the procedure may require early transfusion
  - With ballooning or stenting, vagal stimulation may cause bradycardia, and may need to be treated with atropine
- Complications of the procedure are similar to those of cardiac catheterization
  - Arrhythmias – SVT most common, may see second- or third-degree heart block, sinus bradycardia
  - Bleeding at vascular access sites - appropriate
  - Perforation of cardiac chambers or great vessels by catheters
  - Vascular dissection or hematoma
  - Embolic phenomena – appropriate heparinization may mitigate some of the morbidity

Emergence:

- Infiltration of local anesthetic at vascular access sites limits postprocedure discomfort
- Small amounts of opioid such as fentanyl (1 to 2 μg/kg intravenously) or midazolam may be given to provide postprocedure sedation, thus allowing the patient to remain still and avoid bleeding complications at the femoral vascular access sites

DISPOSITION & MONITORING

- Post-op monitoring may require high intensity (NICU or PICU)
- The alterations in LV systolic function frequently associated with a neonatal presentation usually resolve after relief of the obstruction

COMPLICATIONS

- Immediate postoperative complications:
  - Paradoxical HTN - intravenous administration of nitroprusside with or without esmolol effectively controls the systemic blood pressure during the early postoperative period; longer acting antihypertensive drugs may be needed if hypertension persists
  - Possible sequelae of a bicuspid aortic valve (infective endocarditis and aortic regurgitation)
  - Paraplegia (reflecting ischemic damage to the spinal cord during aortic cross-clamping) after surgical resection of coarctation of the aorta is a rare complication (incidence is reported to be 0.5% to 1.5% for coarctation repair)
  - Abdominal pain may occur during the postoperative period and is presumably due to sudden increases in blood flow to the gastrointestinal tract, leading to increased vasoactivity
- Long-term complications:
  - Persistent/recurrent systemic HTN
  - Residual or recurrent aortic arch obstruction
  - LVH may persist in some children after repair, particularly in those undergoing interventions later in childhood
  - Aortic aneurysms can occur around the area of coarctation or elsewhere in the aorta after surgical intervention or balloon angioplasty
  - Accelerated atherosclerosis and CAD

- Reduced life-expectancy

PATHOPHYSIOLOGY

- Coarctation of the aorta ccounts for ~5% of all congenital heart disease
- Characterized by narrowing (discrete or diffuse) of the descending aortic lumen in the thoracic region
  - **Preductal**: narrowing is proximal to the ductus arteriosus and left subclavian artery
    - If severe, blood flow to the aorta distal to the narrowing is dependent on a patent ductus arteriosus (minimal collateral circulation below the coarctation)
    - Preductal coarctation occurs less commonly and is most likely to present in neonates
  - **Ductal**: narrowing occurs at the insertion of the ductus arteriosus
    - Usually becomes apparent when the ductus closes
    - Usually have well-developed collaterals that supply the area below the coarctation
  - **Postductal**: narrowing is distal to the insertion of the ductus arteriosus
    - More common than pre-ductal type, and is most likely to manifest in older children or young adults
    - Usually have well-developed collateral vessels that supply the area below the coarctation (the spinal cord is supplied by these collaterals)
- Obstruction to aortic flow results and may range from severe obstruction with compromised distal systemic perfusion to mild upper extremity hypertension as the only manifestation
  - Pathophysiologic problems in severe neonatal coarctation include:
    1. Profound left ventricular failure
    2. Impaired coronary perfusion with an increased incidence of ventricular ectopy
    3. Systemic hypotension
    4. PDA-dependent systemic circulation
    5. Systemic hypoxemia
  - Apart from the most extreme variants that become evident in the neonatal period, infants and children with outflow obstruction manifest the effective compensatory mechanisms for pressure overload, often remaining clinically asymptomatic for many years
    - In older children, arterial hypertension is usually found proximal to the region of aortic obstruction and some degree of LVH is present, but ventricular function is usually well preserved
    - Collateral circulation develops with a long-standing pathologic process.
- Repair at an early age is advocated in view of the lower surgical risk in the younger age group and to minimize late morbidity, particularly because this is related to the late development of hypertension
- Complications of coarctation of the aorta include systemic hypertension, left ventricular failure, aortic dissection, premature ischemic heart disease presumably related to chronic hypertension, infective endocarditis, and cerebral vascular accidents due to rupture of intracerebral aneurysms

CONSIDERATIONS IN THE PREGNANT PATIENT

- Coarctation of the aorta represents a fixed obstruction to the forward ejection of left ventricular stroke volume
  - Increases in cardiac output can be achieved primarily by increasing the heart rate
  - During periods of high demand, as during labor or acute increases in intravascular fluid volume produced by uterine contractions, the heart rate may not be able to increase to the extent necessary to maintain adequate cardiac output
  - This sequence of events may result in acute left ventricular failure
- Management of anesthesia:
  - Contractility- maintain
  - Rate- maintain or increase hr
  - Rhythm- NSR
  - Afterload- maintain SVR
  - Preload- maintain
- Analgesia for labor and vaginal delivery is often provided using systemic medications or inhalation analgesia and pudendal block
- General anesthesia is recommended for cesarean section (avoid drop in SVR with regional)
- In all cases, invasive monitoring of arterial and cardiac filling pressures is helpful
- Because the hemodynamic changes seen during labor and delivery can persist into the postpartum period, invasive cardiac monitoring should be continued for 48 hours after delivery in these patients
- Another hazard during labor and vaginal delivery is damage to the vascular wall of the aorta
  - Specifically, with the increased heart rate and myocardial contractility that accompany the pain of labor, the rate of ejection of blood from the left ventricle increases and may lead to dissection of the aorta

REFERENCES

- Coexisting Chapter 3 and 23
- Cote Chapter 15, 16, 21
- Barash Chapter 43
- Miller Chapter 62, 83, 84